

# Lentiginosis and left atrial myxoma

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*A patient is described with cutaneous lentiginosis and left atrial myxoma, an association not previously described.*

The familial association of cutaneous lentiginosis and hypertrophic obstructive cardiomyopathy has recently been recognized. We report a patient with cutaneous lentiginosis and left atrial myxoma, a combination not previously reported. The lentigines were symmetrical and widespread with heavy facial involvement, and were very numerous. The patient's mentality, growth, and sexual development appeared normal.

## Case history

The patient, a clerk aged 18, presented in a dramatic fashion with loss of consciousness and left hemiparesis developing soon after a fist blow on the right cheek.

He was admitted to the neurosurgical unit where bilateral carotid angiography showed complete occlusion of the right middle cerebral trunk 1.5 cm after its origin. Despite anticoagulants and measures to control cerebral oedema, he became deeply unconscious with decerebrate spasms and bilateral upgoing toes. At this stage right lateral craniotomy and removal of bone flap was done to allow decompression. The brain was under tremendous tension but luckily there was no bleeding and after this he gradually improved.

Six days after operation he had a right-sided fit and phenytoin was started. The bone flap was replaced 4 weeks after its removal.

He gave an additional history of transient left-sided weakness 6 months before this event, and later at convalescence suffered an attack of abdominal pain and painful lumps on the fingers. On a single occasion at this time it was noted that he had an apical systolic murmur.

## Family history

His mother, 'heavily freckled', died of heart disease in her mid-forties. His only maternal uncle, similarly pig-

mented, is well. His father, and numerous half sibs are unaffected.

## Examination

A redheaded, fairskinned young man, with generalized

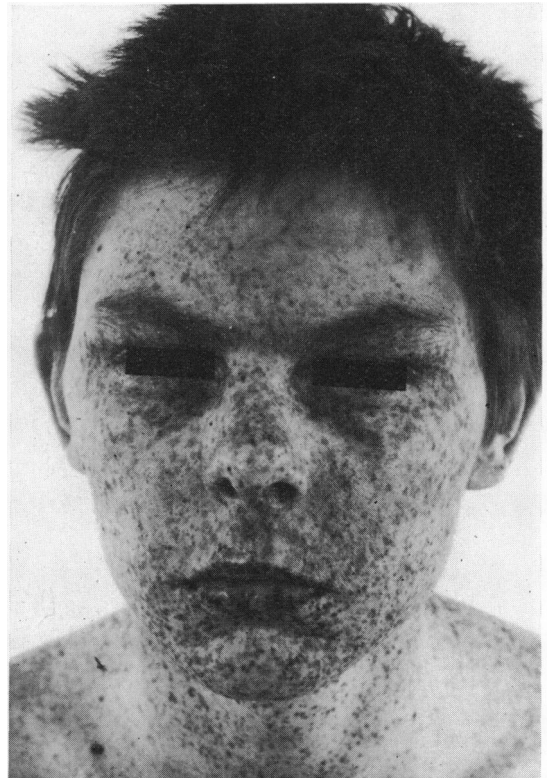


FIG. 1 *Lentigines on face.*

lentiginosis, particularly abundant on the face (Fig. 1), lips, and hands, including the ventral aspects of palms and fingers. The lesions were mostly dark brown but some very dark spots were seen. He was of normal build and height and his sexual development was normal. He showed an intermittent low fever (37°C).

Over 8 days of observation, repeated examination revealed no clinical abnormalities in the heart and vascular system. No murmurs, added sounds, or tumour click were heard at this time. Blood pressure 120/70 mmHg. There was a resolving left spastic hemiparesis, with moderate residual motor disability and normal mental function.

### Investigations

**Blood count** Hb 14 g/100 ml. Red and white cells normal. ESR 40 mm/1st hour.

**Biochemistry** Blood urea, electrolytes, liver function, and aspartate aminotransferase normal. Twenty-four-hour urinary catecholamine excretion normal.

**Skin biopsy** Typical benign lentigo with patchy overactivity of the basal melanocytes without their proliferation. The small extra rete pegs so often seen in lentigo were also present.

**Chest x-ray** Normal.

**Electrocardiogram** Normal.

**Echocardiography** The findings (Fig. 2) were normal movement of the anterior cusp of the mitral valve except for some abnormal forward displacement of the

cusp during midsystole, and multiple echoes typical of left atrial myxoma behind the anterior cusp.

**Right heart catheterization and biplane angiocardiology** Normal pressures throughout, both resting and on exercise. Angiocardiology showed a rounded mobile filling defect in the lower right anterior part of the left atrium.

### Management

Because of the association of lentiginosis, fever, raised ESR, and systemic embolism, hypertrophic cardiomyopathy complicated by endocarditis was first suspected. The true diagnosis was shown by echocardiography, confirmed by angiocardiology, and surgery was done without delay.

### Operation

The patient was explored via a median sternotomy and operation conducted using full cardiopulmonary bypass at normal temperatures using a disposable disc oxygenator. The heart was electrically fibrillated, using a low voltage direct current and the aorta cross clamped. These precautions were taken to avoid the loss of tumour into the systemic circulation. The left atrium was opened exposing an irregular, friable, and gelatinous tumour 6 cm in diameter which had a firm attachment to the inferior margin of the right superior pulmonary vein. The tumour was clearly mobile from this attachment and it was removed entirely from its attachment and its

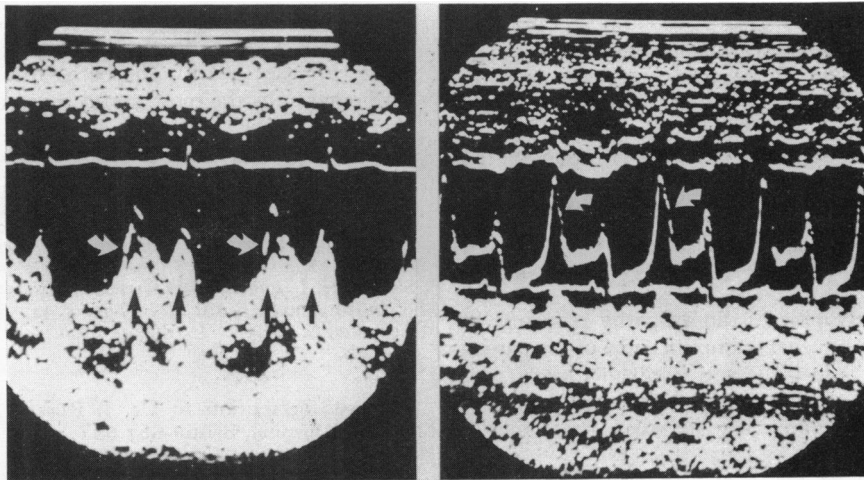


FIG. 2 Preoperative echocardiogram on left. Echo from the anterior cusp of mitral valve (white arrows), and the echoes due to the left atrial myxoma (black arrows) behind it. Post-operative echocardiogram on right. The diastolic slope (white arrows) of the mitral valve echo is steeper than before operation and the echoes behind it have disappeared after removal of the myxoma.

base diathermied. Recovery was uneventful and the patient was discharged home.

### Tumour histology

Typical endotheliomyxoma.

### Follow-up echocardiography

Echocardiography was repeated during convalescence from operation (Fig. 2). The multiple echoes shown behind the anterior cusp before operation had disappeared. The diastolic slope was steeper and the forward displacement of the anterior cusp during midsystole was no longer evident.

### Discussion

Though superficially the lentigo may resemble the freckle, it is very different. It is unaffected by light and has a distinctive histology. A few or many scattered lentigines are to be found in most people, but when they are present in exceptionally large numbers or in a distinctive distribution the condition is named lentiginosis.

Familial lentiginosis was reviewed by Touraine (1955). There is a centrifacial type associated with confluence of the eyebrows, high arched palate, spina bifida, and mental retardation. A second type, associated with gastrointestinal polyposis, has a distribution of lentigines mainly around the orifices, and a string of eponyms too long and disputed to mention. Both are inherited through an autosomal dominant gene.

More recently, a distinctive familial variety has been linked with a cardiac anomaly (Polani and Moynahan, 1972; Somerville and Bonham-Carter, 1972). The spots, symmetrical and widespread, are present in infancy and become increasingly numerous in childhood. In some patients the face is spared. The cardiac lesion in all cases was hypertrophic obstructive cardiomyopathy, and the term progressive cardiomyopathic lentiginosis has been proposed for the syndrome by Polani and Moynahan.

The hypertrophic cardiomyopathy is mainly left sided, but an unusual feature in some of the patients reported by Somerville and Bonham-Carter (1972) has been early right-sided involvement suggesting a diagnosis of pulmonary stenosis. There may be

associated growth retardation, delayed sexual maturity, and at times slight intellectual impairment.

Our patient shows that further syndromes of lentiginosis and cardiac disease may be defined. In him, the lentigines were similar in their apparent familial incidence and increasing development during childhood. They were widespread and, like the patients of Polani and Moynahan (1972), there was a heavy facial involvement. In progressive cardiac lentiginosis they considered two possible links between the cutaneous and cardiac anomalies. One hypothesis is based on the possibility that tumours of neural crest origin are secretory, and that pressor amines released from the lentigines result in the cardiomyopathy. The second alternative, which they favour, suggests a common biochemical anomaly of genetic origin. In our patient, there was no detectable abnormality of catecholamine secretion, and the latter hypothesis could apply, since neural crest material gives rise to the walls of blood vessels, thus permitting a common origin and genetic anomaly for the lentigines and the myxoma. There was some speculation before operation that an unusual tumour might be found based on this premise, but in the event the myxoma was typical in every way.

It has been suggested that patients with lentiginosis should be assessed regularly to detect the presence or subsequent development of cardiomyopathy. Though the heart may be apparently normal, the cardiologist should also think of myxoma. More generally, cardiological assessment may also be rewarding in patients with unexplained systemic arterial obstruction, despite an absence of abnormal signs in the heart. In both instances echocardiography is the most useful screening examination.

### References

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